

## **Cystic Fibrosis- Some Common Problems.**

### **The lungs - coughs and sputum**

People with cystic fibrosis make thick sticky secretions throughout the body. In the lungs the secretion (sputum) obstructs the airways causing them to swell and stretch (bronchiectasis). The airways are then more likely to become infected and damaged.

Many infections like pseudomonas may never go away completely but can be controlled with antibiotics whenever they flare up. At times like this you will usually produce larger amounts of sputum which may taste or smell unpleasant.

Just as important as antibiotics is clearing of the sputum by physiotherapy. Various methods are used to increase sputum clearance these include:

- The active cycle of breathing techniques
- Autogenic drainage
- Flutter
- PEP mask
- Percussion
- Postural drainage
- Coughing

Whichever method is used physiotherapy should be carried out as prescribed. Many patients are helped by taking nebulised bronchodilators which open the airways or DNase which makes the sputum thinner.

### **Exercise**

People with CF can still exercise. This is important but exercise is not a replacement for physiotherapy. It will help improve chest clearance, strength and endurance. Your physiotherapist will discuss this with you.

Please inform us if you:

- Are becoming breathless or more wheezy than usual even if you only notice it when you are exercising.
- Produce more sputum or if it becomes thicker, darker or there is more blood than usual. This can indicate a worsening of your lung infection.

We may be able to help by giving you:

- Intensive physiotherapy
- Antibiotics
- Steroids
- Breathing tablets
- Oxygen

As far as possible we will avoid admitting you to hospital, but sometimes a few days admission will be necessary.

### Common problems

#### Allergic Bronchopulmonary Aspergillosis (ABPA)

*Aspergillus fumigatus* is a fungal infection that grows naturally in the environment. The fungus causes a range of diseases in the human lung. The Growth of *Aspergillus fumigatus* in patients with Cystic Fibrosis is relatively common because of the thick sputum.

#### What is ABPA?

Allergic Bronchopulmonary Aspergillosis is an allergic reaction to the fungus called *Aspergillus fumigatus*. The signs of ABPA is wheezing, which does not improve with the use of bronchodilators. It is sometimes difficult to diagnose ABPA in patients with Cystic Fibrosis because the signs and symptoms are similar to other chest problems.

#### Treatment

Allergic Bronchopulmonary Aspergillosis usually responds to steroids and occasionally to a drug called Itraconazole. The symptoms may flare up from time to time

#### Haemoptysis

Haemoptysis is the coughing up of blood. The changes which occur in the lungs of most people with CF include some stretching of the air spaces and thinning of the lining. They become fragile and can bleed if inflamed, for example during infections. This can be worrying. If mild streaking occurs inform the CF team.

**If heavier bleeding occurs e.g. more than 100mls over 24 hours come to the CF Centre immediately.**

Sometimes the bleeding can be very heavy and may need an operation to prevent it. This operation can be performed in the X-ray or Radiology Department under a local anaesthetic. A fine tube is inserted into the blood vessel in the lungs and is used to block (embolise) the leaking vessel in the lungs. Occasionally if this does not work another operation may be necessary.

#### Nose and Sinuses

Inflammation tends to affect all airways not just in the lungs but also in the windpipe, the throat and the sinuses. This can cause localised swellings called polyps to develop in the nose, a runny nose called rhinitis or blockage of the sinuses which can be painful (sinusitis). Inflammation usually responds to simple treatments like steroid sprays or drops applied to the nose, but sometimes need a specialist ear nose and throat surgeon to investigate the problem.

#### Sex Drugs and Rock and Roll

CF puts a great deal of responsibility on you and may cause varying degrees of emotional stress. The CF team tries to understand this and will not impose unnecessary restrictions on you. Sometimes people experiment with things to bring pleasure or temporary relief from stress. However some of these such as smoking, taking drugs, drinking too much alcohol and sex can also be damaging to the body.

**The CF team will not take a moral attitude or judge you.**

If you have any problems you may wish to talk to your family or friends, rather than 'sweeping them under the carpet'. We are always happy to discuss any problems with you in confidence, either over the phone or face to face.

### **Contact Us:**

If you are concerned about any of the points raised in this leaflet or have any further queries please speak to your consultant or the CF team.

### **Additional Information Sources:**

For more information on all aspects of living with Cystic Fibrosis you can visit the Cystic Fibrosis Trust web site at: [www.cftrust.org.uk](http://www.cftrust.org.uk)

For local news and events the West Midlands Regional Cystic Fibrosis Unit based at Heartland Hospital has its own website at: [www.heartlandscf.org.uk](http://www.heartlandscf.org.uk)

### **Our commitment to confidentiality**

We keep personal and clinical information about you to ensure you receive appropriate care and treatment. Everyone working in the NHS has a legal duty to keep information about you confidential.

We will always ask you for your consent if we need to use information that identifies you. We will share information with other parts of the NHS to support your healthcare needs, and we will inform your GP of your progress unless you ask us not to. You can help us by pointing out any information in your records which is wrong or needs updating.

**Please use the space below to write down any questions you may want to ask:**